A Case of Congenital Coronary-Pulmonary Fistula Involving Both the Conus Artery and the Left Coronary Artery

AUTHOR(S):

NAKAGAWA, AKITO; SAKAMOTO, KIYOSHI; MIKAJIMA, TAKATOSHI; OHSHIMA, TOSHIMI; AKAGI, MASANOXU; HORIO, YUTAKA; OKUMURA, KEN

CITATION:

NAKAGAWA, AKITO ...[et al]. A Case of Congenital Coronary-Pulmonary Fistula Involving Both the Conus Artery and the Left Coronary Artery. 日本外科宝函 1983, 52(4): 565-569

ISSUE DATE:

1983-07-01

URL:

http://hdl.handle.net/2433/208866

Arch Jpn Chir 52(4). 565～569. Juli, 1983

症　　例

A Case of Congenital Coronary-Pulmonary Fistula Involving Both the Conus Artery and the Left Coronary Artery

Akito Nakagawa, Kiyoshi Sakamoto, Takatoshi Mikajima
Toshimi Ohshima, Masanoxu Akagi

The Second Department of Surgery, Kumamoto University Medical School, Kumamoto
(Director: Prof. Dr. Masanobu Akagi)

Yutaka Horio, Ken Okumura

The First Department of Internal Medicine, Kumamoto University Medical School, Kumamoto
(Director: Prof. Dr. Yoshiro Araki)
Received for Publication, April 20. 1983.

Introduction

Following the advent of the modern diagnostic angiography, numerous coronary arterial anomalies have been described. Among them, the congenital coronary arterial fistula is found at the rate of 0.2 to 0.5% of the congenital heart diseases[1], and the coronary-pulmonary fistula forms about 20% of the coronary arterial fistula[2,3,4]. The bilateral coronary-pulmonary fistula forms 5 to 18% of the coronary-pulmonary fistula[3,5]. Although the bilateral anomalies include various types of fistula, the bilateral coronary-pulmonary fistulas involving the conus, the left coronary and the pulmonary artery are very rare.

The subjective symptoms of fistula are influenced by the age, shuntratio, and the types and positions of the abnormal vessels. Most of the patients do not have any symptom until they develop general fatigue, palpitation, shortness of breath, exertional dyspnea and unconsciousness as a result of steal phenomenon. The heart murmur which leads to the discovery of coronary pulmonary fistula varies case by case. Therefore, angiography is needed to make an accurate diagnosis for a type of coronary-pulmonary fistula involving both the conus artery and the left coronary artery.

The operative methods contain the ligation of the fistula or closure by suturing of it from intracavity of PA with the use of the extracorporeal bypass.

Key words: Conus artery, Left coronary-pulmonary fistula, Continuous murmur, Steal phenomenon, Closure of the fistula.
索引語：円錐動脈，左冠動脈肺動脈瘻，連続性雑音，スチール現象，瘻孔閉鎖.
Present address: The Second Department of Surgery, Kumamoto University Medical School 1-1-1. Honjo, Kumamoto 860 Japan.

Case Report

A 16 years old male with headache, vertigo, nausea, vomiting and unconsciousness was admitted to our hospital in October, 1976. The patient had never experienced such symptom previously.

Pulse rate was 82. Blood pressure was 150/60-0 mmHg. Laboratory findings including the complete blood count, liver, kidney and lung functions, bleeding time and clotting time were normal. There were no abnormalities on inspection and palpation. On auscultation of the chest a grade 2/6 to and fro heart murmur was heard highest at the second sound. Chest x-ray showed cardiac-thoracic ratio 40%, normal pulmonary markings and slight prominence of the left pulmonary arc. ECG revealed neither LVH nor ST-T change in sinus rhythm. Phonocardiogram showed heart murmur of to and fro type, which was markedly increased and became almost continuous by pharmacological (Methoxamine HCL: Mexan) and exertional overload test. Echocardiogram showed no abnormality.

The heart catheteri ation revealed the left to right shunt of 12% in the pulmonary artery which was further increased by Mexan overload test. The selective coronary arteriogram showed that the conus artery originating from aorta just above the orifice of the right coronary artery passed around the anterior side of the aorta and the posterior side of PA to flow into the angiomatous bulge on the anterior wall of PA (Fig. 1 AB). Simultaneously the small abnormal channels from the left coronary artery were seen to drain into the bulge. (Fig. 1C).

The heart was exposed through median sternotomy. The angiomatous bulge of about 0.5 cm in diameter was found on the anterior wall of PA, into which several small vessels were seen to flow (Fig. 2AB). In addition, the tortuous conus artery of 2 mm in diameter was seen to

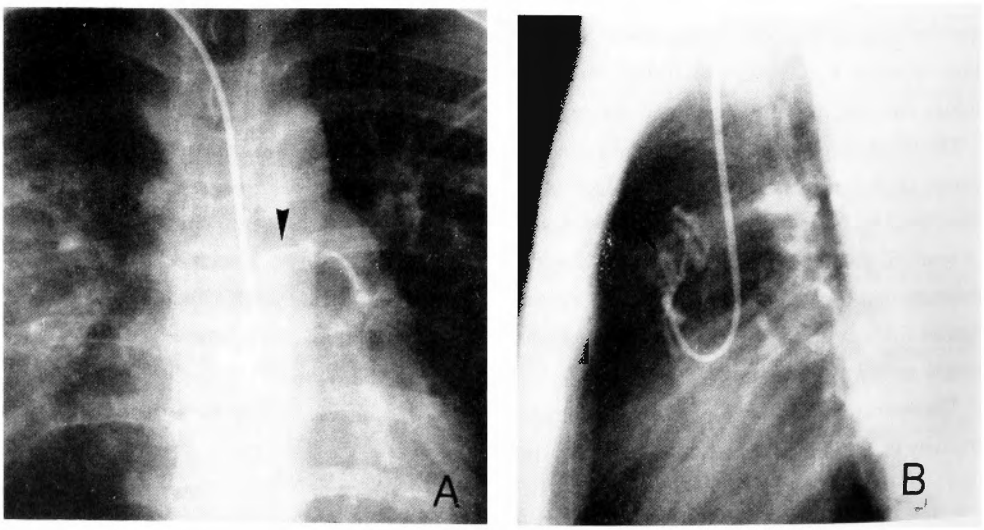

Fig. 1 AB. Selective right conal arteriogram showed that the conal artery (arrow) originated from just above the right coronary artery to flow into PA, after passing around the anterior side of the aorta and the posterior side of PA.
(A, Anterior-posterior view and B, Lateral view).

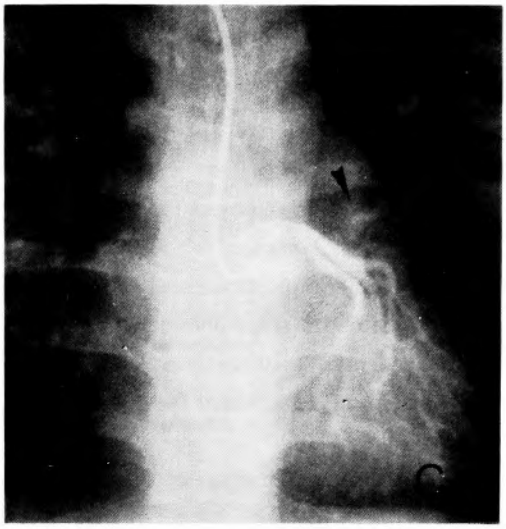

Fig. 1 C. Antero-posterior view of the left coronary arteriogram. The small abnormal channels (arrow) from the left coronary artery were seen to drain into PA.

flow into the bulge around the left side of PA, together with other abnormal small vessels originating from the coronary artery. The ligations of the conus artery and some other small vessels were performed successfully one by one with Ti-cron threads.

Discussion

On the mechanism of origin of the congenital coronary pulmonary fistula, GOBEL described that the abnormal vessels might be arterial in origin based on histology and they might have originated from PA and connected with the coronary artery from the aorta.[6] On the contrary,

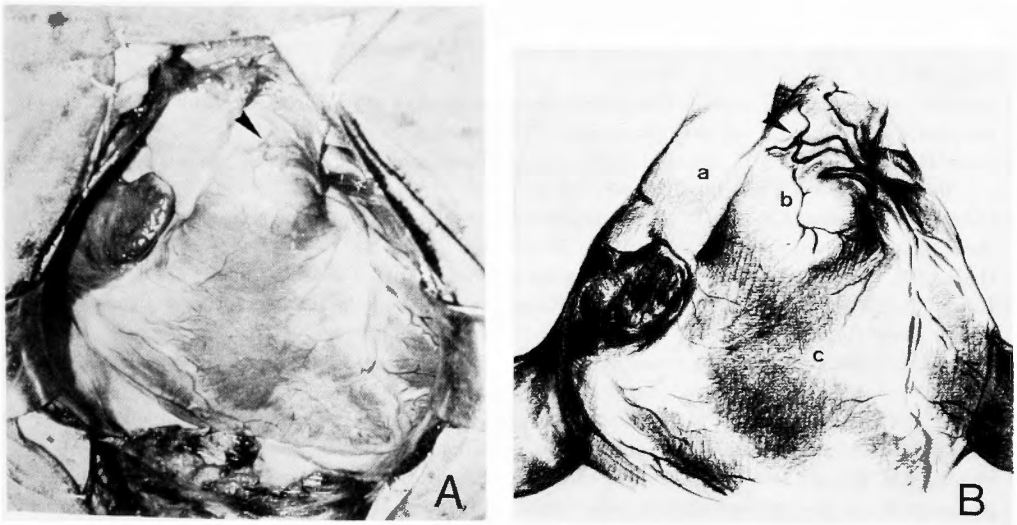

Fig. 2. Operative view, A and its diagram, B showing the angiomatous bulge (arrow) on the anterior wall of PA, into which several tortuous small vessels were seen to flow. a; Aorta,b; Pulmonary trunk, c; Right ventricle

OGDEN et al. proposed that the abnormal vessels might be venous in histologically and they might be a remnant of the arterio-sinusoidal vessels[7]. Also, ERNST proposed that the abnormal vessels might be connected to the vasa-vasorum of PA[8,9]. However, all of the above mentioned theories are neither completely nor adequately substantiated. Thus the mechanism of its formation is remained to be clarified.

According to OLDHAM[3], the subjective symtoms are influenced by the age, shunt-ratio and the types and positions of the abnormal vessels, although most patients have no subjective complaints. Patients have a variety of complaint such as shortness of breath, exertional dyspnea and unconsciousness once they obtain the steal phenomenon. Furthermore, sometimes these patients have severe heart diseases such as ischemic or congestive heart disease, coronary insufficiency, infectious endocarditis, valvular disease (papillary muscular dysfunction) and unconsciousness.

The heart murmur is one of the most common objective symptoms of this disease, that is mainly continuous or to and fro type which are heard in most cases according to OLDHAM[3], LEVIN[2], and HENRY[4]. Differentiation of many heart diseases which exhibit the same type of heart murmur is difficult to achieve and one must be very careful to make a diagnosis when the heart murmur of continuous or to and fro type is obtained.

The operative method advocated for this anomaly contains either ligation of the fistula as in our case or closure by suturing of it from intracavity of PA using the extracorporeal circulation[10,11].

References

1) Krongrad E, Ritter DG, et al: Pulmonary atresia or severe stenosis and coronary artery-to-pulmonary artery fistula. Circulation 1972; **46**: 1005–12.
2) Levin DC, Fellow KE, et al: Hemodynamically significant primary anomalies of coronary arteries. Angiographic aspects. Circulation 1978; **58**: 25–32.
3) Oldham HN, Ebert PA, et al: Surgical manatement of congenital coronary artery fistula. Ann Thorac Surg 1971; **12**: 503–14.
4) Neufeld HN, Lester RG, et al: Congenital communication of a coronary artery with a cardiac chamber or the pulmonary trunk. ("coronary artery fistula") Circulation 1961; **24**: 171–9.
5) Baim DS, Kline H, et al: Bilateral coronary arterypulmonary artery fistulas. Report of five cases and of the literature. Circulation 1982; **65**: 810–5.
6) Gobel FL, Anderson CF et al: Shunts between the coronary and pulmonary arteries with normal origin of the coronary arteries. Am J Cardiol 1970; **25**: 655–61.
7) Ogden JA, Congenital anomalies of the coronary arteries. Am J Cardiol 1970; **25**: 474–9.
8) Ernst CB, Kassen KP, et al: Vascular malformation overlying the pulmonary artery simulating a patent ductus artery.
9) Tobin CE. Some observations concerning the pulmonic vasa vasorum. Surg Gynecol Obstet 1960; **111**: 297–303.
10) Cooley DA, Hallman GL, et al: Definitive surgical treatment of anomalous origin of left coronary artery from pulmonary artery. Indication and Results. J Thorac Cardiovasc Surg 1966; **52**: 798–808.
11) Symbas PN, Schlant RC, et al: Congenital fistula of right coronary artery to right ventricle complicated by Actinobacillus a tinomyctemcomitans endarteritis. J Thorac Cardiovasc Surg 1967; **53**: 379–84.

和文抄録

円錐動脈左冠動脈—肺動脈瘻の1治験例

熊本大学医学部第二外科（主任：赤木正信教授）
中川　昭十，酒本喜与志，三ケ島尊利，大嶋　寿海，赤木　正信

熊本大学医学部第一外科（主任：荒木淑郎教授）
堀尾　　豊，奥村　　謙

両冠動脈—肺動脈瘻は非常に稀なものであるが，われわれはその1種である右円錐動脈左冠動脈—肺動脈瘻の1例を，結紮により治癒せしめたので報告する．患者は16才の高校生の男子．生来健康であったが，高校入学後特に疲れ易くなってきたので精査のため，本学第一内科に入院してきた．胸部聴診で，第2肋間胸食左縁に Levine Ⅱ度の to and fro の雑音があり，心カテの結果肺動脈レベルで左右短絡があることが証明された．最初，PDA を疑ったが，撰択的冠動脈造影により，両側冠動脈肺動脈瘻の診断がつき，手術目的にて当科に転科してきた．手術は胸骨正中切開で行い心臓に達すると，肺動脈前壁に指示頭大の血管腫様腫瘤があり，周りから異常血管が6～7本流入していた．その異常血管を1本ずつ2—3重結紮して，瘻孔閉鎖を行い，手術を終わった．術後は，雑音も消失し，経過良好であった．